Indication of Kasai's Operation for Biliary Atresia : For Early Decision of Liver Transplantation

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Summary

To determine the surgical indication for KASAI's operation for biliary atresia is to consider the indication of liver transplantation. Determination of the liver transplantation before KASAI's operation is comparatively difficult. We devised a scoring system from the history and the laboratory examinations, and drew ellipses for confidences of histological calculation to determine the surgical indication of this disease.

After the KASAI's operationè indication of the liver transplantation is determined by their clinical course. Some of the type of obstruction of extrahepatic bile duct showed poor prognosis, and some of the type of variation of the vessels had tendency of poor prognosis. Postoperative cholangitis and the change of the laboratory examinations have important role in persistent jaundice. To make the early decision for indication of liver transplantation, careful and meticulous early postoperative observation must be done.

Introduction

KASAI's operation has taken the most important role for salvage the annoying disease, biliary atresia. However, one-fourth of the patients still showing poor prognosis. From our experience, some of them have no bile excretion after the operation, and some of them showed persistent jaundice in spite of the bile excretion. These are the most adequate patients for liver transplantation. In Japan, we have a tendency to operate for all patients without consideration of the indication for surgery except for the older patients. This indiscriminate surgery sometimes leads to difficulties in performing the second surgery, i.e., liver transplantation. In this report, we studied the adequate indication for surgery for biliary atresia, and focused on the indication of the liver transplantation before and after the KASAI's operation.

Subjects and Methods

Two hundred and fourty-one patients were operated upon in the Tohoku University Hospital

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during a period of 34 years from 1953 to July 1987. One hundred and one patients are now living with or without jaundice. Using these clinical materials, we have reported on their prognosis[1~5]. This paper is a summary of these reports to determine surgical indication for biliary atresia and for liver transplantation.

Results

1. *Indication for KASAI's operation*:

Table 1 shows the relationship between the age at the operation and postoperative bile excretion in our recent 145 cases. In the patients receiving surgery before the age of 60 days, good bile excretion rate was 91%, while the rate was below 45% for patients receiving surgery after the age of 91 days. In patients older than 121 days, we selected the surgical cases according to their clinical and histological findings.

A review of the data showed that most of the results of laboratory examinations varied greatly among the groups with different prognosis. Therefore, we devised a scoring system

Table 1. Biliary atresia: Age at operation and bile excretion

Age at operation	No. pts	Bile excretion		
		Good	Poor	None
— 60 days	55	50(91%)	3	2
61— 70	33	23(70)	8	2
71— 90	38	26(68)	8	4
91—120	14	6(43)	2	6
121—	5	3(60)	2	0
Total	145	108(74)	23	14

Table 2. Score table for determination of the surgical indication of biliary atresia

Items of examination	Group		
	A	B	C
Age at operation (days)	≦70	105～140	140<
Alpha-2 globulin (%)	—	—	17<
Beta globulin (%)	<13	15<	—
Gamma globulin (%)	<11	11～20	20<
CCFT	—	++	+++ or ++++
ZTT (units)	≦ 4	5～15	15<
TTT (units)	≦ 3	5～10	10<
GOT (units)	≦250	250～400	400<
GPT (units)	≦160	300～360	360<
Alkaline phosphatase (KA units)	—	—	100<
Total cholesterol (mg/100 ml)	<300	—	—
Albumin (%)	55<	45～55	<45
Birth weight (g)	—	<2500	—
Score	+1	−1	−3

from the history and the laboratory examinations (Table 2). We chose 13 items of examination for this table and divided the data of each item into 3 groups; group A (excellent), group B (intermediate), and group C (poor). Each group comprizes 10 of the 13 items. Seventy nine patients were analyzed by this scoring system, and 19 of them showed minus scores. These 19 patients had no active bile excretion after the operation. Recently we changed a little: unit of the alkaline phosphatase changed from King Armstrong unit to International Unit and if it shows more than 1200 IU/L, it belongs to group C. The patients having a minus score are considered as requiring liver transplantation instead of KASAI's operation.

Prognosis of the patient may be predicted by histological findings of the liver specimen which was taken by needle biopsy. Fig. 1 shows the relationship between the ratio of the percentage of bile duct proliferation to the intrahepatic artery and age at operation[2]. The ellipse shows the confidence of 95% level of the cured cases. Postoperative bile flow may be poor, if the ratio falls outside the ellipse. The same tendency was seen in the relationship between the ratio of the percentage of bile duct proliferation to the intrahepatic portal vein and age at operation (Fig. 2). From these clinical and histological factors, we can establish the surgical indication

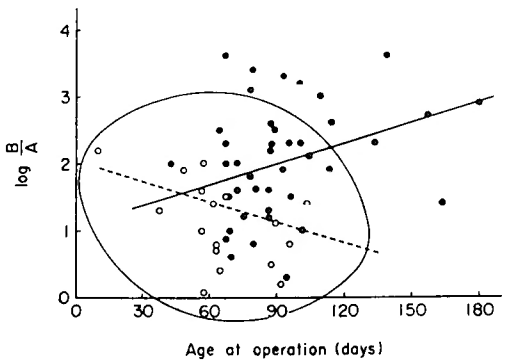

Fig. 1. Relationship between the ratio of the percentage of bile duct proliferation to the intrahepatic artery and age at operation. The ellipse shows the confidence of 95% level of the cured cases (open circle).

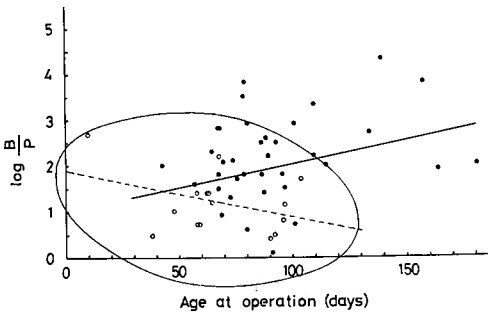

Fig. 2. Relationship between the ratio of the percentage of the bile duct proliferation to the intrahepatic portal vein and age at operation. The ellipse shows the range of confidence at 95% level of the cured cases (open circle).

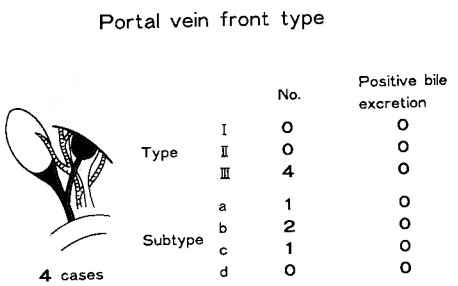

Fig. 3. Portal vein front type

The biliary system at the forefront, followed by the portal vein and the hepatic artery was found in 4 patients. All showed poor bile excretion.

for biliary atresia. Indication for the liver transplantation can be determined in some patients before the KASAI's operation.

2. *Indication of liver transplantation by operative findings*:

We may anticipate the prognosis by the type of the obstruction of the bile duct. However, it is difficult to choose the patient for liver transplantation. Only III-c type (obstructed at the porta hepatis and absence of the common bile duct) is considered to be the indication of the liver transplantation. Poor prognosis may be predicted by the type of the arrangement of the vessels in the vicinity of the porta hepatis[6]. Portal vein front type has usually bad results (Fig. 3).

3. *Post operative course and indication for the liver transplantation*:

Re-operation was carried on 36 patients. Among them, 23 showed good bile excretion, once, after the first operation. The results of re-operation depends on the bile excretion after the primary operation. For the patients who showed good bile excretion after the primary operation, good bile excretion was seen in 91% after re-operation, while in the patients who showed no bile excretion, bile excretion rate was 54% and only half of them survived. Therefore, the remaining patients may be considered to be candidates for transplantation.

The most important factor to improve the operative results is to overcome cholangitis. Among patients who survived for more than 1 year after the operation, 18 patients have jaundice. Persistent jaundice without a history of jaundice was seen in 5 patients. These patients may be candidates for liver transplantation. The remaining 13 patients experienced a reduction of the bilirubin level to normal range, previously. Jaundice recurred and continued after they suffered from cholangitis. Patients whose bilirubin level exceeded 10 mg/dl showed bad prognosis. These patients are also good indication for transplantation.

To investigate the clinical factors for pejsistent jaundice, we analysed the laboratory data.

Table 3. Biliary Atresia: Rate of changes of examination data during 3 months after primary operation

No. of cases		Persistent Jaundice Bilirubin 5 mg/dl< 10	Persistent Jaundice 2～5 mg/dl 8	Jaundice free cholangitis (+) 8	Jaundice free (−) 8
Al-P	elevated	80%	62.5%	75%	62.5%
	unchanged	20	37.5	25	25
	declined	0	0	0	12.5
γ-GTP	elevated	87.5	100	50	0
	unchanged	12.5	0	25	50
	declined	0	0	25	50
TTT	elevated	78	100	37.5	0
	unchanged	22	0	37.5	50
	declined	0	0	25	50
γ-gl	elevated	100	67	14.3	0
	unchanged	0	33	85.7	62.5
	declined	0	0	0	37.5

Table 3 shows the rate of variation in 4 laboratory examinations (alkaline phosphatase, gamma-GTP, TTT, and gamma-globulin) for 18 patients with jaundice and 16 patients without jaundice. Among patients with jaundice, many patients showed a tendency of increase in each parameter. We can predict their prognosis within 3 months after the operation.

Comment

To determine the surgical indication for liver transplantation in biliary atresia before the KASAI's operation is not easy. Many factors should be considered to determine the indication. In biliary atresia, abnormality of the liver function and/or histological deterioration is progressive with the age. Therefore, the age (days) at operation is an important factor in this disease. From our experience, the upper limit of the successful operation is around 140 days. The prognosis of the patients with extremely abnormal liver function tests and/or histological deterioration are bad. For these patients KASAI's operation should not be done. Patients with minus score by our scoring system[1], and/or the patients whose histological calculated ratio falls outside the ellipse which already described above[2] are the most adequate candidates for liver transplantation. However, donors of the liver in infants is not so much and in the country that the liver transplantation is not allowed generally, KASAI's operation is also undertaken for them. Even scanty bile drainage can improve the abnormal condition, and it may lengthen their lives, but some of them died shortly after the operation.

Surgical indication for liver transplantation is mostly applied to the patients who showed continuous jaundice even after the KASAI's operation or to the patients who showed persistent jaundice after disappearance of jaundice. MURAJI and his co-workers[7] reported that the liver transplantation should be considered for those with recurrent jaundice with constant increase of serum bilirubin exceeding 10 mg/dl. And they add that those with an absence of initial bile drainage or with deterioration of hepatic disorder at 6 months after operation are candidates of liver transplantation. The liver function and the histological structure will be deteriorated by time in patients with mild jaundice even if their bilirubin is below 10 mg/dl. Esophageal varices, hypersplenism, and other complications will also appear. Therefore, care should be taken for these patients whose bilirubin level is between 5 to 10 mg/dl. Careful postoperative observation is important for early decision of the liver transplantation. Elevation of the data of alkaline phosphatase, gamma-GTP, TTT and gamma-globulin in the early postoperative days suggests the unconvenient complication, such as persistent jaundice.

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和文抄録

胆道閉鎖症に対する葛西手術の適応
——肝臓移植の早期の決定のために——

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胆道閉鎖症の手術適応に関する報告は少ない．本邦においては日令がきわめて遅い例以外は手術されるのが普通である．しかし，手術を施しても胆汁がでない例や，黄疸が消褪しない例があり，このような例では初めから肝移植の適応になる可能性がある．ここではこのような例を見分けるために臨床的事項や検査値を用いてスコアリングするシステムを紹介し，また，肝生検標本の計測による適応決定のための棄却楕円図を紹介した．

一方，手術後の患者の移植の適応のために手術所見のみならず，術後早期の検査値の変動，特にアルカリフォスファターゼ，ガンマ GTP, TTT, ガンマグロブリンなどの変動をみることが重要で，これらが上昇して来る場合には黄疸が持続したり，合併症が起こり易く，改善しない場合には移植を含めた対策をたてるべきである．